

# Fitting Free-Form Scleral Lenses on a Patient with Limbal Stem Cell Deficiency Secondary to Stevens-Johnson Syndrome

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## BACKGROUND

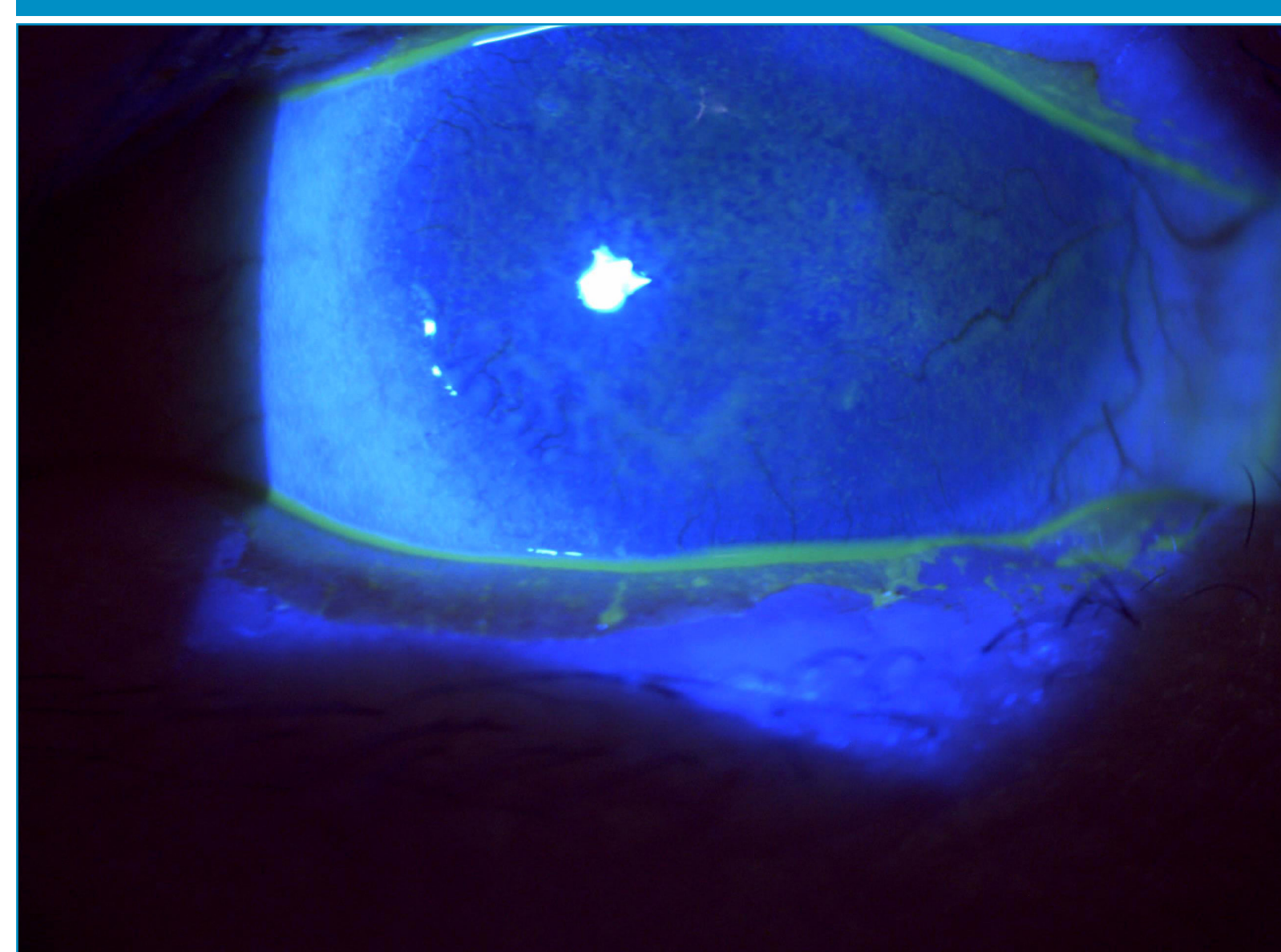
Limbal Stem Cell Deficiency (LSCD), a chronic ocular sequelae seen in patients with Stevens-Johnson Syndrome (SJS), involves the destruction of limbal stem cells caused by chronic ocular surface inflammation and replacement of the corneal epithelium by abnormal conjunctival tissue.<sup>1</sup> SJS is characterized by the destruction of epithelium and mucus membranes.<sup>2</sup> Scleral lenses (SL) may be used as an option to optimize the health of the ocular surface.<sup>3</sup> As new SL technology emerges, it is worthwhile to examine how profilometry-driven SL increase the efficiency of the fitting process while providing optimal comfort with lens wear.<sup>4</sup>

## CASE DESCRIPTION

- 39-year-old Caucasian female presented for dry eyes.
- Previously diagnosed with LSCD secondary to SJS of unknown etiology since 1998.
- OSD managed with Xiidra, preservative free artificial tears, and Tobradex. Medications for the condition are being managed by an outside corneal specialist.
- Current scleral lenses are 3 years old, and patient reports constant cloudy vision. Uses PuriLens for filling solution.
- Entering corrected VA: 20/40 OD, 20/30 OS
- Slit lamp examination of cornea: 360 degrees of peripheral neovascularization and opacification with 4+ diffuse punctate epithelial erosions OU (Figure 1). OS showed neovascularization extending to the visual axis (Figure 2).
- Tomography showed irregular astigmatism with keratometry values of 42.2 / 44.2 D OD, and 40.1 D / 44.2 D OS (Figure 3).
- Due to keratinization of the conjunctiva and irregularity of the corneal surface, a free-form design SL was ordered using the Pentacam CSP software (Figure 4).
- A diagnostic lens was applied to determine the appropriate base curve and power for the initial SL order.
- Over the course of twelve weeks, a total of three lenses were ordered to improve landing zone alignment and corneal vault.

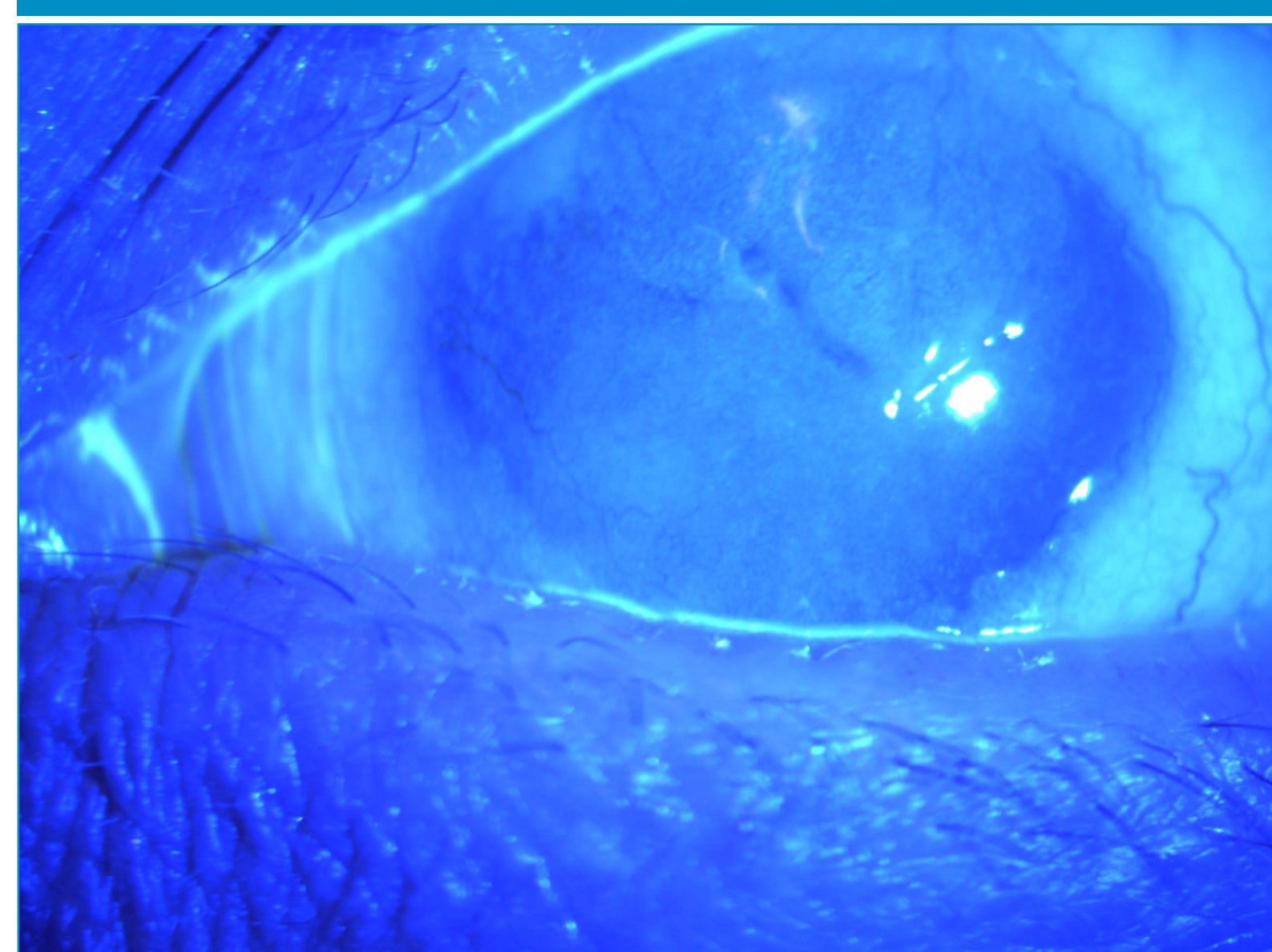
**FIGURE 1**

Slit lamp photo of OD showing neovascularization and diffuse 4+ PEE.



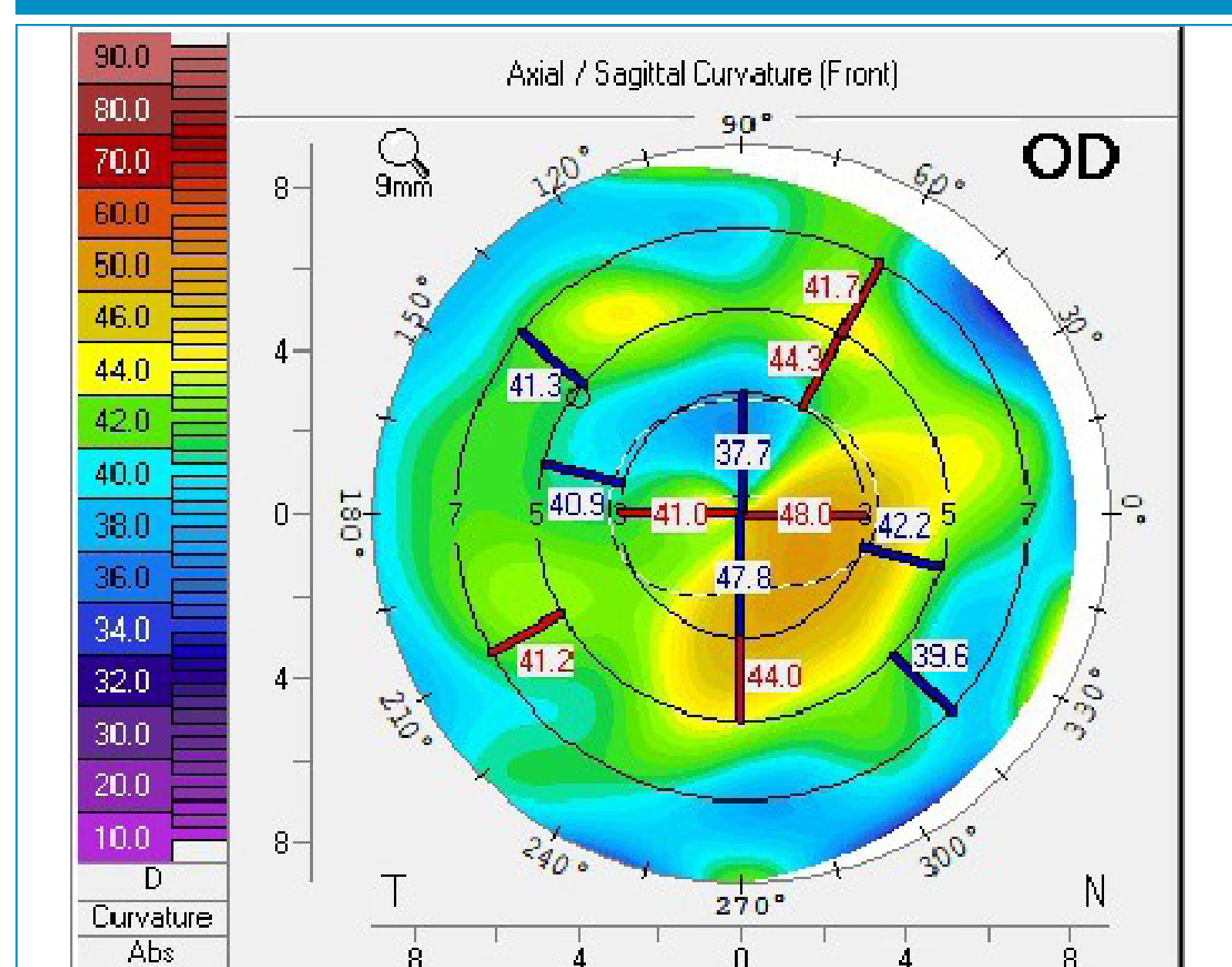
**FIGURE 2**

Slit lamp photo of OS showing neovascularization and diffuse 4+ PEE.



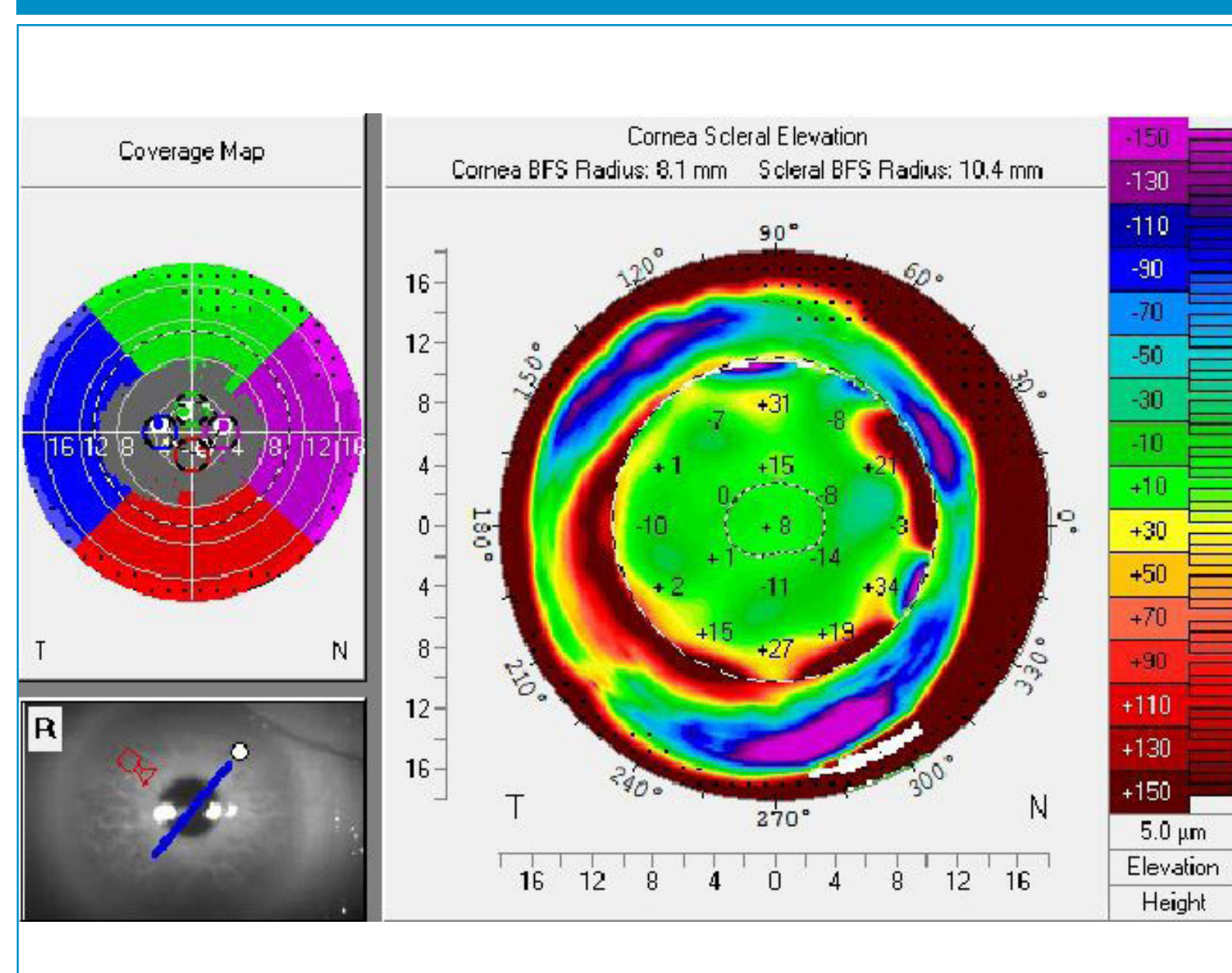
**FIGURE 3**

Tomography of OD illustrating the highly irregular nature of the patient's cornea with areas of steepening and flattening within the pupil.



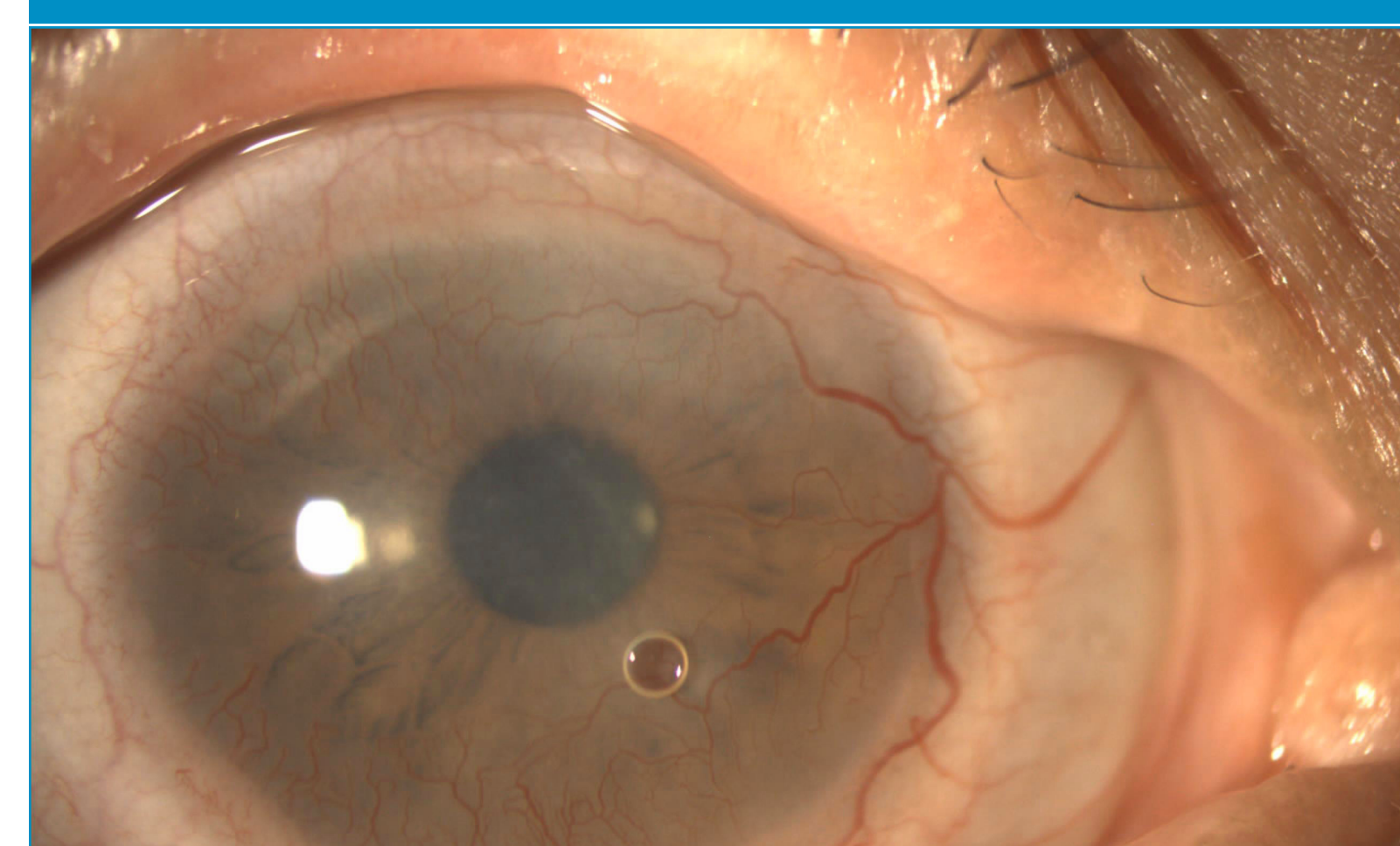
**FIGURE 4**

OD Pentacam CSP Report used to order a free-form design scleral lens showing irregular variations in corneal and scleral elevation.



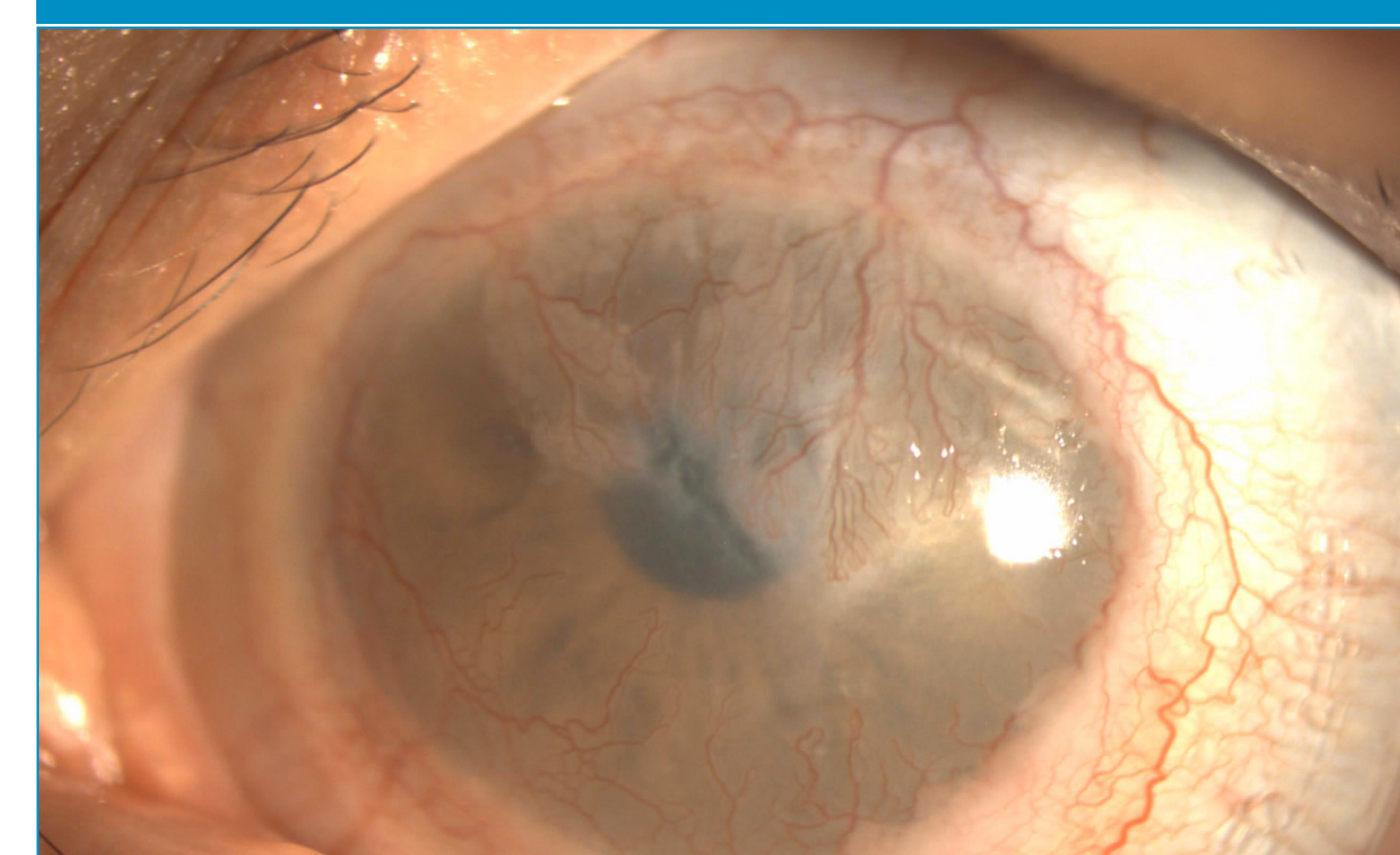
**FIGURE 5**

Final scleral lens fit OD showing adequate clearance over areas of neovascularization, and great edge alignment with mild edge lift nasally. VA: 20/25



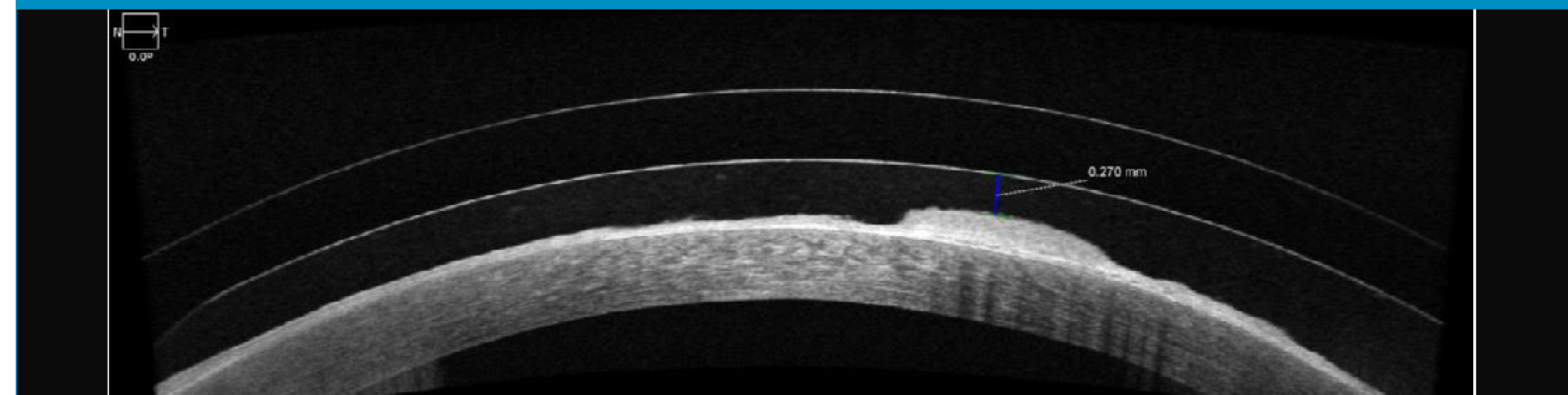
**FIGURE 6**

Final scleral lens fit OS showing adequate clearance over areas of neovascularization, and great edge alignment with mild edge lift nasally. VA: 20/30



**FIGURE 7**

AS-OCT of final lens design OS showing 270  $\mu$ m of clearance over area of neovascularization after 2.5 hours of wear.



**TABLE 1**

Final scleral lens design OD and OS

	Type	Power	BC	Diameter	Total Sag	Edges by Quadrant	Material
OD	BostonSight Smart 360	-5.13	8.32	17.00	2754	1: 0 2: 0 3: 0 4: 0	Optimum Extreme
OS	BostonSight Smart 360	-1.28	8.72	17.00	2813	1: 0 2: 0 3: 0 4: 100	Optimum Extreme

## CONCLUSIONS

- This case illustrates how free-form design scleral lenses have the potential to improve outcomes of patients with severe ocular surface disease.
- Profilometry allowed for exact measurements of the highly irregular cornea and scleral landing zones, evident on the CSP report.
- A diagnostic fitting alone would cause countless challenges with the conjunctival irregularities demonstrated in this case, outlining the versatility of this methodology to improve patient outcome in terms of comfort and wear time.

## REFERENCES

- [1]Choi, S. H., Kim, M. K., & Oh, J. Y. (2019). Corneal Limbal Stem Cell Deficiency in Children with Stevens-Johnson Syndrome. *American journal of ophthalmology*, 199, 1–8. <https://doi.org/10.1016/j.ajo.2018.10.016>
- [2]Sotozono, C., Ueta, M., & Yokoi, N. (2018). Severe dry eye with combined mechanisms is involved in the ocular sequelae of SJS/TEN at the chronic stage. *Investigative Ophthalmology & Visual Science*, 59(14). <https://doi.org/10.1167/iovs.18-24019>
- [3]Yoon, H. H., Harthan, J. S., Nau, A. C., Vogt, J. S., Nau, C. B., Schornack, M., & Shorter, E. (2023). Topographical Characteristics of Patients with Ocular Surface Disease Successfully Fit in Custom Scleral Lenses. *Investigative Ophthalmology & Visual Science*, 64(8).
- [4]Yoon, H., Harthan, J., Skoog, W., Nau, A., Fogt, J., Nau, C., Schornack, M., & Shorter, E. (n.d.). *Process and Outcomes of Fitting Corneoscleral Profilometry-Driven Scleral Lenses for Patients with Ocular Surface Disease*. Poster presented at Global Specialty Lens Symposium (2023).

## CONTACT INFORMATION

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